Nodular Scleroderma: Case Report and Literature Review

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ABSTRACT. Objective. To describe a unique case of scleroderma (SSc) presenting as multiple keloidal nodules and early-onset osteoarthritis (OA), and to summarize the clinical and serological data for 13 similar patients reported in the English literature since 1966.

Methods. MEDLINE review of the literature over a 35-year period (1966-2002) revealed 13 cases of nodular SSc. We describe a case of nodular SSc in a 40-year-old African-American male with localized SSc who developed progressive skin thickening and keloidal nodules on the arms, hands, chest, abdomen, and thighs with advanced osteoarthritis of the hips.

Results. In all 14 cases, diagnosis was made based on skin biopsy and evidence of keloid (nodule) formation. Ten cases occurred in women and 4 in men, with ages ranging from 9 to 66 years and a mean age of 38.9 years. The ethnicity of the patients was given in only 5 of the 13 previously reported cases. Including our patient, 4 were of African descent, and 2 were Caucasian. Most patients had symptoms of SSc consisting of arthralgias (n = 10), sclerodactyly (n = 9), Raynaud’s phenomenon (n = 8), digital pitting and/or calcinosis (n = 5), shortness of breath with pulmonary fibrosis (n = 5) or pulmonary hypertension (n = 1), dysphagia or reflux (n = 3), renal disease (n =3), and elevated erythrocyte sedimentation rate (n = 3).

Conclusion. Nodular SSc is a rare variant that presents with lesions that clinically resemble keloids. OA, as documented in the present case, does not appear to be a typical feature of nodular SSc. His family history was negative for connective tissue disease or keloid formation.

Physical examination revealed indurated skin on the forehead, scalp, thorax, arms, and digits. In addition, there were discrete, irregular, hypertrophic plaques resembling keloids localized to the arms and thorax (Figure 1). Salt-and-pepper pigmentary changes of the scalp and forehead were noted. His palmar skin displayed contractures that resulted in atrophy of the thenar and hypothenar skin. He had pain with rotation and flexion of his hips. The remaining components of his examination were unremarkable.

Nodular scleroderma (SSc) is considered a rare variant of the disease1-4 that may occur in the setting of either systemic sclerosis1-7 or localized scleroderma (morphea)8-12. We describe a 40-year-old African-American man with localized SSc presenting as multiple keloidal nodules. Our patient’s disease was complicated by severe early onset osteoarthritis (OA) affecting the hips. Although scleroderma has been associated with rheumatoid arthritis13,14, this is the first reported case of nodular SSc associated with advanced OA. The etiology of nodule formation in SSc is unknown.

MATERIALS AND METHODS

Literature review. A literature review using the MEDLINE key words scleroderma, morphea, nodular, and keloid produced 13 other cases of nodular SSc in the English literature over a 35-year period (1966 to 2002)1-12. All patients exhibited characteristic nodular lesions but the degree of keloid formation varied. Diagnosis of SSc and keloid formation was based on histological confirmation and/or examination by a dermatologist. Clinical and serological data from all 14 cases are summarized in Table 1.

Case report. A 40-year-old African American male with a medical history of hypertension presented after noticing the slow onset of focally thickened skin over the thorax, arms, and fingers. There was no prior history of keloids or abnormal scarring. Review of systems disclosed a 2-year history of worsening hand and knee arthralgias and lower back pain. There was no morning stiffness, oral ulcers, visual changes, dysphagia, reflux, alopecia, dyspnea, or symptoms of Raynaud’s phenomenon. Medications included amiodipine 5 mg daily for hypertension. His family history was negative for connective tissue disease or keloid formation.

Physical examination revealed indurated skin on the forehead, scalp, thorax, arms, and digits. In addition, there were discrete, irregular, hypertrophic plaques resembling keloids localized to the arms and thorax (Figure 1). Salt-and-pepper pigmentary changes of the scalp and forehead were noted. His palmar skin displayed contractures that resulted in atrophy of the thenar and hypothenar skin. He had pain with rotation and flexion of his hips. The remaining components of his examination were unremarkable.

Laboratory evaluation revealed normal complete blood count and differential, liver function tests, chemistry profile, and erythrocyte sedimentation rate, as well as negative antinuclear antibody, extractable nuclear antigen screen, and anti-topoisomerase I (Scl-70) antibodies. Pulmonary function testing, echocardiogram, and nailfold capillary microscopy were normal. Radiographs revealed advanced osteoarthritic changes involving both hips. A biopsy of indurated skin from the inner surface of his right arm showed a minimal superficial perivascular inflammatory cell infiltrate and fibrosis within the dermis consistent with SSc. A later biopsy of skin from the ventral surface of his right arm showed the broad, brightly eosinophilic collagen bundles typical of a keloid.

DISCUSSION

The terms nodular SSc, keloidal SSc, pseudokeloidal SSc, keloid-like morphea, and nodular morphea have all been used to describe patients with features of either localized or systemic SSc who present with hypertrophic plaques that may clinically resemble keloids1-12. Although keloids are...
Table 1. Clinical and serological features of patients with nodular SSc.

<table>
<thead>
<tr>
<th>Author Reference, year</th>
<th>Sex</th>
<th>Age*</th>
<th>Areas of Skin Involvement</th>
<th>Type of Scleroderma</th>
<th>Symptoms</th>
<th>Positive Laboratory Tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>Cantell, 1980</td>
<td>M</td>
<td>66</td>
<td>Trunk, neck, arms</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, renal involvement</td>
<td>BUN 27 mg/dl</td>
</tr>
<tr>
<td>James, 1984</td>
<td>M</td>
<td>24</td>
<td>Arms, thighs</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, pulmonary involvement, esophageal dysfunction</td>
<td>ANA, nucleolar</td>
</tr>
<tr>
<td></td>
<td>F</td>
<td>17</td>
<td>Chest, back</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement</td>
<td>ANA, speckled</td>
</tr>
<tr>
<td>Sasaki, 1992</td>
<td>F</td>
<td>66</td>
<td>Abdomen</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement, esophageal dysfunction</td>
<td>ANA, speckled, RF</td>
</tr>
<tr>
<td>Wilson, 1992</td>
<td>F</td>
<td>42</td>
<td>Chest, abdomen, arms</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement</td>
<td>ANA, speckled</td>
</tr>
<tr>
<td>Yamamoto, 1994</td>
<td>F</td>
<td>36</td>
<td>Chest</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, renal and pulmonary involvement</td>
<td>ANA, speckled, anti-Scl70 ESR, antiRNP, CrCL, 27ml/min</td>
</tr>
<tr>
<td>Krell, 1995</td>
<td>F</td>
<td>40</td>
<td>Trunk, neck, arms</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias</td>
<td>ESR, BUN 41mg/dl, ANA, speckled</td>
</tr>
<tr>
<td>Mizutani, 1995</td>
<td>F</td>
<td>42</td>
<td>Chest</td>
<td>Systemic</td>
<td>Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement</td>
<td>ANA, speckled, anti-Scl70</td>
</tr>
<tr>
<td>Akintewe, 1985</td>
<td>F</td>
<td>17</td>
<td>Trunk, neck, arms</td>
<td>Localized</td>
<td>Sclerodactyly, arthralgias</td>
<td>None</td>
</tr>
<tr>
<td>Micalizzi, 1994</td>
<td>F</td>
<td>64</td>
<td>Arms, breast, abdomen, back</td>
<td>Localized</td>
<td>Esophageal dysfunction</td>
<td>ANA, speckled</td>
</tr>
<tr>
<td>Stephanato, 1992</td>
<td>M</td>
<td>20</td>
<td>Suprascapular, supraclavicular</td>
<td>Localized</td>
<td>None</td>
<td>ESR</td>
</tr>
<tr>
<td>Kubo, 1997</td>
<td>F</td>
<td>61</td>
<td>Neck, chest, back, face</td>
<td>Localized</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>Hsu, 1999</td>
<td>F</td>
<td>9</td>
<td>Right arm, right axilla, right chest</td>
<td>Localized</td>
<td>Arthralgias</td>
<td>Not reported</td>
</tr>
<tr>
<td>Cannick, 2003</td>
<td>M</td>
<td>40</td>
<td>Arms, chest, abdomen, thighs</td>
<td>Localized</td>
<td>Arthralgias</td>
<td>None</td>
</tr>
</tbody>
</table>

* Age of presentation with keloidal nodules. RP: Raynaud’s phenomenon; ANA: antinuclear antibody; RF: rheumatoid factor; BUN: blood urea nitrogen; CrCL: creatinine clearance; ESR: erythrocyte sedimentation rate; RNP: ribonucleoprotein; Scl70: scleroderma 70.

Figure 1. Photograph shows a large keloid on the ventral arm. In addition, there are numerous flat indurated, hyperpigmented plaques on the lateral chest and medial surface of the arm. These lateral lesions are typical of localized scleroderma (morphea).
well-known phenomena primarily in individuals of African
descent, their formation in SSc is rare. In patients with
nodular SSc, keloids may represent an aberrant response to
the early inflammatory phase of the disease\(^2\).

Our case report is similar to other reported cases of
patients with SSc who later developed keloids or nodules.
The distribution of the lesions primarily on the arms and
upper torso in our patient is also similar to other case
reports. However, none of the clinical photographs in the
literature so far show the dramatic keloid formation
observed in our patient.

To date, our patient with nodular SSc is the only one
described with symptoms and radiographic evidence of
advanced OA. It is our opinion that whatever autoimmune
process is driving his SSc is also driving his OA. The lack
of symptoms of esophageal dysmotility, overt Raynaud’s
phenomenon, sclerodactyly, and telangiectasias suggests
that our patient has localized rather than systemic SSc. His
condition progressed rapidly despite treatment with hydrox-
ychloroquine and methotrexate.

Nodular SSc is a rare disorder affecting a minority of
patients with SSc. Comparisons between cases are
confounded by the variability in terminology, presentation,
symptoms, and appearance of the lesions. The pathogenesis
is still undetermined, but may involve a combination of
environmental and genetic factors.

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